**Exercise performance in Shone’s Syndrome and relationship with outcomes.**

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Background

Shone’s syndrome was described first in 1963 as a complex that characterized by severe left heart obstruction. It has 4 known components: supravalvular ring, parachute mitral valve, subaortic stenosis, and coarctation of the aorta. For diagnosis, 3 out of 4 components must be present. Clinical and echocardiographic findings have been well described, but exercise performance in these patients is not well known.

Methods

We reviewed the Adult Congenital database from June 1987 to July 2014 and cross-matched with the Mayo Clinic Stress Center database. We included patients with Shone’s syndrome who had a cardiopulmonary exercise test (CPX) and echocardiogram (echo) at the same visit. We selected the first CPX test for each patient and correlated with the closest echo. We reviewed the Mayo Clinic electronic medical record for clinical and demographics variables and outcomes.

Results

From a database of 6,231 adult congenital patients, we found a total of 6 Shone’s patients with CPX and echo. There were 4 men, 2 women. Age range was 20-45 years. All patients had repaired coarctation of the aorta. Number of cardiac surgeries ranged from 2-8. Ejection fraction by echo was reduced in 2 patients (13% and 18%), otherwise ≥ 50%. Good effort was achieved on the CPX by all patients. Peak VO2 ranged from 38% to 105% predicted. Over follow-up of 7-9 years, 2 patients died and 1 underwent heart transplant. The Table identifies clinical, echo findings, and exercise performance in all patients separated according to unfavorable or favorable outcomes.

Conclusion

Exercise capacity is highly variable in patients with Shone´s syndrome ranging from 38% up to 105%. Patients who died or underwent cardiac transplant not surprisingly had a more complex clinical history with more echo abnormalities, and they also more frequently exhibited poor performance on the CPX.

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| **Table. Demographic, clinical, echocardiographic and Cardiopulmonary stress test variables in patients with Shone's Syndrome** |
| **Patient** | **1** | **2** | **3** | **4** | **5** | **6** |
|  **Outcomes** |
| Death | - | + | + | - | - | - |
| Transplant | + | - | - | - | - | - |
|  **Clinical History** |
| Male | + | + | - | + | - | + |
| Age≥ 40  | - | - | - | + | - | + |
| Surgeries > 4 | + | - | + | - | - | - |
| AVR | - | + | + | - | - | - |
| MVR | + | + | - | + | + | - |
| Heart Failure | + | + | + | - | - | - |
|  **Echocardiographic Results** |
| EF < 50% | + | + | - | - | - | - |
| LV Enlargement | + | + | - | - | - | - |
| RVSP ≥ 50 mmHg | + | + | + | - | + | - |
|  **Cardiopulmonary Exercise Test Results** |
| Peak VO2 < 60% | + | + | + | - | + | - |
| VE/VCO2 nadir > 35 | + | - | + | - | - | - |
| HR Recovery < 13 bpm | + | - | + | - | + | - |

AV = aortic valve replacement; MVR = mitral valve replacement; EF = ejection fraction; RVSP = right ventricle systolic pressure, HR = heart rate.